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CASE REPORT

Prosthetic Rehabilitation of Hypohidrotic Ectodermal Dysplasia: A Case Report

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***ABSTRACT*:**
Ectodermal dysplasia is a rare group of inherited conditions in which two or more ectodermally derived anatomical structures fail to develop or partially develop. Hypohidrotic ectodermal dysplasia is most common type in which patients have sparse scalp and body hair, reduced ability to sweat along with oligodontia or total anodontia leading to loss of function like chewing, speech and affects the appearance of the patient. Prosthodontic management can be completed with removable, fixed, overdenture, or implant-retained prostheses. Early dental treatment can improve the appearance of the patient, which leads to less emotional and psychological problems to the patient. For rehabilitation, it is important to know the age, number and condition of present teeth, and the status of growth of the patient. A 18 year-old male patient who was reported the dental department was treated by removable partial denture in maxillary arch and complete denture in mandibular arch**.**

Key words: Ectodermal dysplasia, Partial denture, Partial anodontia

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INTRODUCTION:

Ectodermal dysplasia is a genetic disorder in which ectodermally derived anatomic structures fail to develop or partially develop. It is manifested as hypohidrosis, hypotrichosis and hypodontia or partial anodontia. Hypohidrotic ectodermal dysplasia (Christ-Siemen Touraine syndrome) is most common type and symptoms include sparse scalp and body hair, reduced ability to sweat along with missing teeth. It may be inherited in an X-linked recessive, autosomal recessive or autosomal dominant manner depending on the genetic cause of the condition. However the most common mode of inheritance is X-linked so male is affected commonly however female patients show partial expression of the abnormal gene, i.e. teeth may be reduced in number or may have mild structural changes. Hypohidrotic ectodermal dysplasia (HED) is caused by mutations in the EDA, EDAR, or EDARADD genes EDA is the only gene known to be associated with X-linked HED (XLHED). Ninety-five percent of individuals with HED have the X-linked form. The genes EDAR and EDARADD are known to be associated with both autosomal dominant and autosomal recessive forms of HED (Bartlett et al., 1972). Mutations in these genes account for 5% of HED (Wright et al., 2013). The EDA, EDAR and EDARADD genes provide instructions for making proteins (ectodysplasin A) that work together during embryonic development. Ectodysplasin A forms a part of a signaling pathway that is critical for the interaction between two cell layers, the ectoderm and the mesoderm. In the early embryo, these cell layers form the basis for many of the body organs and tissues. Ectoderm-mesoderm interactions are essential for the formation of several structures that arise from the ectoderm, including the skin, hair, nails, teeth and sweat glands. Mutations in the EDA, EDAR or EDARADD gene results in defective ectodysplasin A formation thereby preventing normal interactions between the ectoderm and the mesoderm and hence impairing the normal development of hair, sweat glands and teeth. The improper formation of these ectodermal structures leads to the characteristic features of hypohidrotic ectodermal dysplasia.

Occurrence rate is 1-7 in 10000–100000 births. The mortality rate is 30 percent in infancy or early childhood because of intermittent hyperpyrexia. Patients show heat intolerance and pyrexia due to reduced sweat glands.

Hypoplasia or aplasia of skin, hair, teeth, nails, reduced density of eyebrow & eyelash hair, periorbital wrinkling of skin and hyper pigmentation are another symptoms associated with HED. Affected individual also have characteristic facial and oral abnormalities including prominent forehead, sunken nasal bridge (saddle nose), midface hypoplasia etc. The skin on most of body parts may be abnormally thin, dry and soft appearing as prematurely aged appearance. Reduced salivary gland development leads to varying degree of xerostomia. In some of the cases decreased function of certain components of immune system i.e. decreased lymphocytic function, cellular immune hypofunction causing increased susceptibility of certain infection and allergic condition is seen. Oral presentation of such individual usually have hypodontia, anodontia, teeth if present is peg shaped or conical in shape. Alveolar ridge development is poor. Treatment of hypohidrotic ectodermal dysplasia may include special hair care formulas or wigs, measures to prevent overheating, removal of ear and nose concretions, and dental evaluations and treatment e.g., restorations, dental implants, or dentures (Prasad et al.2012 and Bajaj 2015).

CASE REPORT:

A 18 years old boy reported to dental dept. of Nalanda Medical College and Hospital, Patna; complaining of missing teeth and masticatory difficulty. He was accompanied by his father who gave the history of missing teeth since childhood except two anterior and four posterior teeth of upper jaw. He also gave the history of fever since childhood and he was intolerant to heat and takes frequent dips in summer to keep cool. There was no family history of missing teeth and other features. Extraoral examination revealed the typical features of HED like frontal bossing, sparse scalp hair, missing eyelashes and eyebrows, saddle nose, protuberant and everted lips (Fig.1&2). The boy was moderately built and fairly nourished. His skin was dry and the body hair was scanty.

Intra Oral and Radiographic Examination Results

Mandibular arch was completely edentulous with poorly developed alveolar ridge and the maxillary arch had two standing central incisor, conical in shape and two molars in each side of the arch. Radiographic investigation confirm that there is no impacted teeth anywhere in the arch. (Fig.3)

Diagnosis:

Since hypohidrosis, hypotrichosis and hypodontia were evident on examination, the boy was diagnosed with Hypohidrotic Ectodermal Dysplasia (HED) with partial anodontia.

Prosthodontic management

The prosothodontic management of a patient suffering from HED depends on degree of anodontia/hypodontia. In case of complete anodontia, the treatment would comprise of complete denture either conventional or implant supported one. In patient with partial anodontia removable/fixed partial dentures and over denture may be considerd.

Prosthodontic Management in present case:

A removable partial denture in maxillary arch and complete denture in mandibular arch were planned**.**Maxillary central incisors (conical in shape) were modified to receive PFM crown to improve the esthetic of the patient (Fig.4). Preliminary impression were made using alginate (ZELGAN,DENTSPLY, INDIA, Fig.5). Custom trays were prepared using auto polymerising acrylic resin(DPI-RR COLD CURE,DPI,MUMBAI,INDIA) after giving spacer to mid platine raphe, incisive papilla and crest of the mandibular ridge. Border moulding was done using green stick compound (DPI MUMBAI,INDIA). Secondary impression were made using ZOE impression paste(DPI,MUMBAI,INDIA). Record bases were made on the master cast to establish maxillomandibular relation and then they were mounted on articulator. The maxillomandibular relation was based on fullness of lower face, appearance of lips, height of lower face compared to upper freeway space and phonetic test.

Teeth were selected in accord to dentogenic concept and keeping alveolar ridge in mind and while arranging the teeth basic denture guidelines for teeth placement were used.

Trial dentures were checked for retention, occlusion, phonetics and esthetics. Trial denture were processed using heat cure acrylic resin (DPI) and after polishing denture were inserted. The boy was taught about insertion and removal of dentures and was given post insertion instruction on hygiene and maintenance and he was restricted to soft diets initially (Fig. 9).Patient was very happy and confident after rehabilitation.

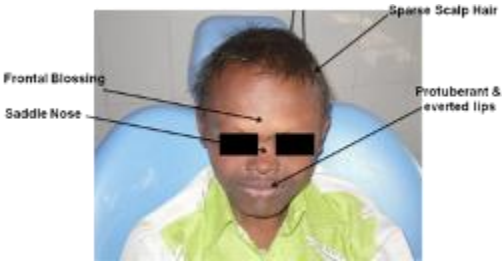

Fig. 1: Frontal view of Patient: Extra-oral examination revealed the typical features of HED like frontal bossing, sparse scalp hair, saddle nose, protuberant and everted lips.

Fig. 2: Lateral view of Patient

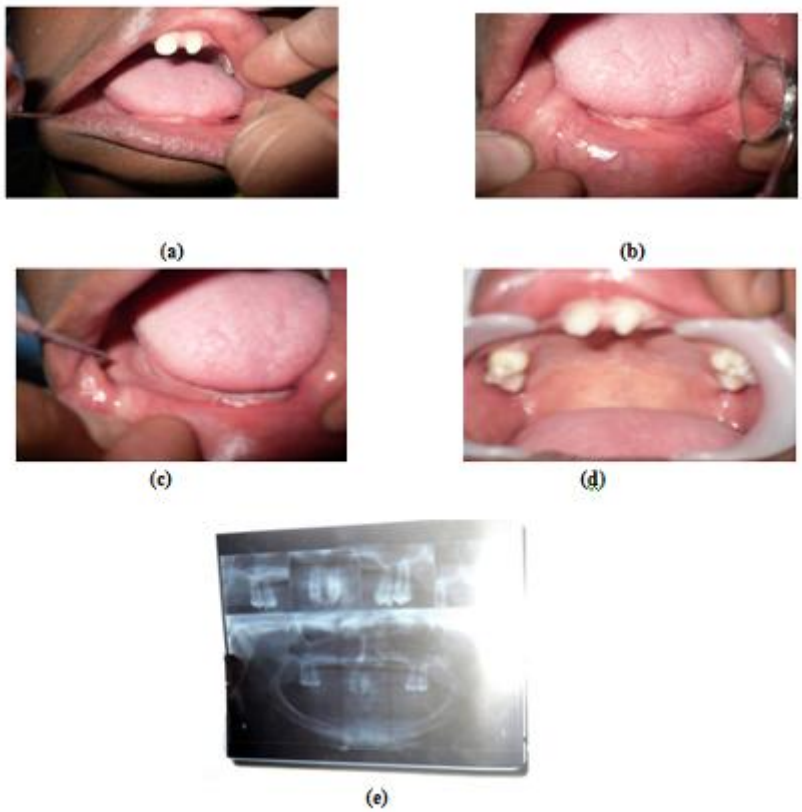

Fig. 3: Intraoral view of Patient
a) overall intraoral view: dry mucosa, large and fissured tongue and anterior conical crowns.
b) Mandibular view: Poorly developed Mandibular alveolar ridge
c) Mandibular view: Poorly developed whole mandibular ridge and macrogollsia due to long period of edentulousnes
d) Maxillar view: Poorly developed maxillary alveolar ridge, conical shaped anterior crown & reduced diameter of molar crowns as well as oligodontia.
e)OPG: Poorly developed maxillary & mandibular alveolar ridge and Hypodontia

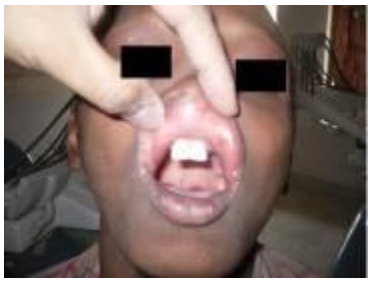

Fig. 4: Modification of conical shaped crown by placing metal fused ceramic jacket crown.

Fig. 5 a: Jaw record taken after verifying CR and VDO b: Wax occlousal rim of upper and lower arch

Fig. 6: Final impression of the patient

Fig 7: Upper and lower arch master cast

Fig. 8 RPD of upper arch with clasp given to improve retention and stability and complete denture of lower arch

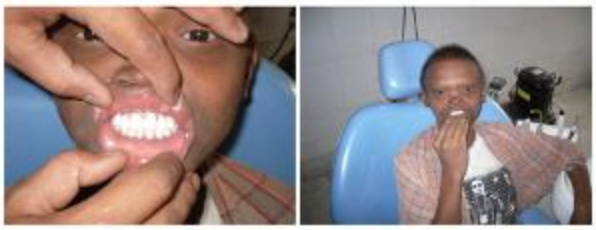

Fig. 9 Trial of dentures for retention, occlusion, phonetics and esthetics.

Fig. 10: Rehabilitation: Patient wearing prosthesis and feeling confidence

DISCUSSION

There is little information available in the literature regarding treatment modalities of a young patient having HED. In case of this patient, the diagnosis was done at the age of 6 when he suffered from repeated bouts of fever that required medical attention whereas he was reported to dental OPD at the age of 16 years. Management of such a case requires psychological attention too. The most common dental treatment in cases of ED is removable complete/partial dentures where as FPDs and Implant supported prosthesis are to be considered when seemed feasible. Patients with ED present a relatively thin and under developed residual ridge covered by thin mucosa. These features along with the decreased quantity of saliva are the main concern encountered in treating such cases. In present case dentures were made flat and the amount of saliva was sufficient for the dentures to adhere. There is no definite time to begin treatment, but it is recommended that initial prosthesis could be delivered when the child starts schooling, so that child he/she could have a better appearance and have time to adapt to the prosthesis.

Masticatory difficulty is a major problem in such patient due to absence of teeth however with dentures in mouth improvement in masticatory function as well as esthetics were observed in the patient due to which there was a boost in self confidence of the patient. A removable partial denture/conventional complete denture is often a suitable treatment choice because of the need to easily modify the prosthesis when required also the treatment modality is easy, economical and suitable for the patient. Periodic recalls of a patient with ED is quite important because of prosthesis adjustment when needed. The treatment goals for this patient were to establish a functional occlusion with prosthetic rehabilitation and to obtain a esthetic smile.

CONCLUSION

Prosthetic management of a patient with HED is important. It provides esthetics, phonetics and masticatory comfort. It maintains healthy supporting tissue. It boost the patient self confidence.

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